Prenatal diagnosis of right ventricular outflow tract obstruction with intact ventricular septum, and detection of ventriculocoronary connections

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Abstract

Objectives—To determine the accuracy of prenatal diagnosis of pulmonary atresia and intact ventricular septum (PAIVS), and pulmonary stenosis, including prenatal detection of ventriculocoronary connections, to evaluate heart size during the prenatal period, and to evaluate the outcome.

Design and patients—Medical records of 20 cases with prenatally diagnosed PAIVS and pulmonary stenosis were reviewed retrospectively. Prenatal and postnatal echocardiography were also reviewed and dimensions of the ventricles and vessels were measured retrespectively.

Results—Of 20 prenatal diagnoses (15 PAIVS and five pulmonary stenosis), 16 were confirmed as correct. One critical pulmonary stenosis case had been diagnosed as PAIVS prenatally; three had no confirmation. Eight pregnancies were terminated, three had no active treatment, and nine were treated; all survived. Of 13 assessed with ventriculocoronary connections prenatally, seven were diagnosed correctly (four with, three without ventriculocoronary connections), but one was falsely positive; five had no confirmation. The more prominent hypoplasia of the main pulmonary artery and the tricuspid valve annulus, and the sigmoid shape of the ductus arteriosus, seemed to be associated with the presence of ventriculocoronary connections.

Conclusions—Current prenatal echocardiography can accurately diagnose right ventricular outflow tract obstruction and ventriculocoronary connections. Prenatal detection of this constellation of abnormalities aids in family counselling and decisions on postnatal management.
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Keywords: congenital heart disease; pulmonary atresia; ultrasonography; prenatal diagnosis

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Right ventricular outflow tract obstruction with an intact ventricular septum is a major cardiac lesion that presents during the neonatal period. Because it is ductus dependent, several interventions are required soon after birth.[1] Some neonates have additional severe morphological abnormalities, such as a hypoplastic right ventricle or ventriculocoronary connections, which limit strategies for surgical or catheter intervention.[2–4] Prenatal diagnosis of right ventricular outflow tract obstruction and intact ventricular septum has been documented, but little information is available as to the pathogenesis of the severe form.[5–8]

More recently, high resolution echocardiography has made it possible to assess precise structures in the human fetus from the third month; even ventriculocoronary connections can be detected prenatally.[9] Concomitantly, several studies on fetal cardiac intervention, which can improve right ventricular growth in cases of right ventricular outflow tract obstruction and intact ventricular septum, have been published.[10–12] It has therefore become much more important that fetal echocardiographic findings be accurate, not only for the major diagnosis but also for the presence of ventriculocoronary connections, which can affect outcome of prenatal intervention.

In this study we retrospectively reviewed all records of prenatal diagnoses of pulmonary atresia and intact ventricular septum (PAIVS) and pulmonary stenosis available at our paediatric centre. Using these data, we determined the accuracy of prenatal diagnosis, including prenatal detection of ventriculocoronary connections, evaluated the heart size during the prenatal period, and evaluated outcomes.

Method

FETAL CASES

We identified all prenatal diagnoses of PAIVS or pulmonary stenosis recorded at the Hospital for Sick Children, Toronto, Ontario, Canada, between January 1989 and December 1997. Fetuses diagnosed with right ventricular outflow tract obstruction associated with more complex heart disease were excluded. Patients with severe tricuspid regurgitation, such as occurs in cases of tricuspid dysplasia and Ebstein's anomaly, were excluded.

EQUIPMENT

Prenatal echocardiography was performed using Advanced Technology Laboratories (Bothell, Washington, USA) equipment— either an Ultramark 9 system with a 5 MHz curved array probe, or a high density imaging system with a 7–4 or 4–2 MHz curved array probe. Postnatal echocardiography was performed with a 5 MHz or a 7–4 MHz phased array probe (Advanced Technology Laboratories), or with a Hewlett-Packard (Andover, Massachusetts, USA) Sonos 1500 or 2500 system with a 7.5 or 5.5 MHz phased array probe.

All images were recorded on videotape for offline analysis.

ACCURACY OF PRENATAL DIAGNOSIS AND OUTCOME

All prenatal and postnatal records were reviewed to determine the accuracy of the prenatal echocardiographic diagnosis of PAIVS or pulmonary stenosis (judged against the corresponding records of cardiac catheterisation and necropsy data), and to evaluate the prenatal and postnatal course.

PRENATAL DETECTION OF VENTRICULOCORONARY CONNECTIONS

Since February 1994, when we first observed ventriculocoronary connections in a fetus with PAIVS during prenatal echocardiography, we attempted to examine the presence or absence of ventriculocoronary connections in all cases with a prenatal diagnosis of PAIVS or severe pulmonary stenosis. The systematic evaluation to assess ventriculocoronary connections involved scanning the aortic root and right ventricular myocardium using colour Doppler mapping with a velocity setting of 32 cm/s or less. When turbulent signals were identified in the myocardium, we determined the direction of the flow using the pulsed Doppler interrogation. In general, flow in the coronary arteries was identified by the presence of forward diastolic and reversed systolic flow, whereas an intracavity right ventricular connection had a turbulent diastolic pattern.[1 9 13] Hence, presence or absence of ventriculocoronary connections was decided at the time of fetal echocardiography. In the current study, all scans were reviewed, and prenatal ventriculocoronary connections findings were retrospec-

tively categorised as "obvious", "suspicious", or "absent". "Obvious" indicated a typical coronary Doppler flow pattern of systolic high velocity retrograde flow and diastolic low velocity antegrade flow. "Suspicious" indicated a turbulent colour signal around the right ventricle, but without a typical Doppler flow pattern. Accuracy of the prenatal diagnosis of the presence or absence of ventriculocoronary connections was later assessed by postnatal right ventriculography and necropsy reports, and if present, ventriculocoronary connections were described therein as small or large.

MEASUREMENTS OF HEART SIZE

Serial measurements were made from prenatal and postnatal echocardiograms. Four dimensions were defined for consistent measurement[14]: diameters of the tricuspid valve annulus and the mitral valve annulus, measured in a four chamber view immediately before closure of the atrioventricular valves; the diameter of the ascending aorta above the aortic root; and the diameter of the main pulmonary artery midway between the pulmonary valve and the bifurcation. All vessel measurements were noted at their maximum dimensions, presumably at the end of systole. These diameters from both prenatal and postnatal echocardiography were compared to normal fetal heart measurements from the Children's Hospital, Boston, Massachusetts.[14]

In addition, the shape of the ductus arteriosus was assessed arbitrarily. The shape of the ductus arteriosus was described as: sigmoid, if the ductus had a long meandering course and joined the left pulmonary artery; or straight, if it arose from the aortic arch at an acute angle from the normal position and had a short

Table 1 *Prenatal and postnatal anatomy of fetuses with right heart obstruction with intact ventricular septum, and outcome*

| Case | GA | Reason(s) for referral | Main diagnosis | | Diagnosis of VCC | | Shape of DA | Outcome* |
			Prenatal	Confirmed	Prenatal	Confirmed		
1	33	Small RV	PAIVS	PAIVS	na	Large	Sigmoid	Status: Fontan (7 years)
2	19	Small RV	PAIVS	—	Obvious	—	Unclear	Aborted
3	34	Small RV	PAIVS	PAIVS	Obvious	Large	Sigmoid	Status: BCPS (3 years)
4	31	Small RV, twin	PAIVS	PAIVS	Obvious	Large	Sigmoid	Status: BCPS (2 years)
5	20	Small RV	PAIVS	PAIVS, SAS, bicuspid AoV	Obvious	Large†	Straight	Aborted; necropsy
6	19	Small RV	PAIVS	PAIVS	Obvious	—	Sigmoid	Aborted; necropsy
7	30	Small RV	PAIVS	PAIVS	Suspicious	Small	Straight	Status: BCPS (2 years)
8	38	Dandy-Walker	PAIVS	PAIVS	na	—	Unclear	Death
9	36	Small RV	PAIVS	PAIVS	Absent	Absent	Straight	Status: BCPS (2 years)
10	30	Small RV	PAIVS	PAIVS	Suspicious	Absent	Straight	Status: BT (1 year)
11	21	Small RV	PAIVS	PAIVS	na	—	Straight	Aborted; necropsy
12	24	Small RV	PAIVS	—	na	—	Straight	Aborted
13	17	Small RV	PAIVS	PAIVS	na	—	Straight	Aborted; dysmorphic fetus, by necropsy
14	22	Thickened RV	PAIVS	—	Absent	—	Straight	Aborted
15	24	Small RV	PAIVS	PAIVS	Absent	—	Sigmoid	Aborted
16	38	Small RV, family history	PAIVS	Severe duct dependent PS	na	Small	Straight	4p–deletion; death
17	27	Severe PS, family history	Severe PS	Severe duct dependent PS	Absent	Absent	Straight	Status: balloon valvuloplasty (1 year)
18	22	TTTS	Severe PS	Severe duct dependent PS	Absent	—	Straight	Ischaemic changes in brain; death
19	32	IDDM, previous stillborn	Severe PS	Moderate PS	na	Absent	Straight	Status: balloon valvuloplasty (6 years)
20	31	Previous stillborn (21 trisomy)	Severe PS	Moderate PS, ASD	Absent	Absent	Straight	Status: ASD closure, valvuloplasty (2 years)

AoV, aortic valve; ASD, atrial septal defect; BCPS, bidirectional cavopulmonary shunt; BT, Blalock-Taussig shunt; DA, ductus arteriosus; GA, gestational age (in weeks) at initial study; IDDM, insulin dependent diabetes mellitus; na, not assessed; PAIVS, pulmonary atresia and intact ventricular septum; PS, pulmonary stenosis; RV, right ventricle; SAS, subaortic stenosis; TTTS, twin to twin transfusion syndrome; VCC, ventriculocoronary connections.
*Status is the patient's postoperative status (at patient's age). Death in all cases occurred after no active treatment (by parental request).
†Probably large, as suggested by the presence of a dilated tortuous coronary artery found during necropsy. All other ventriculocoronary connections confirmations were by postnatal right ventriculography.
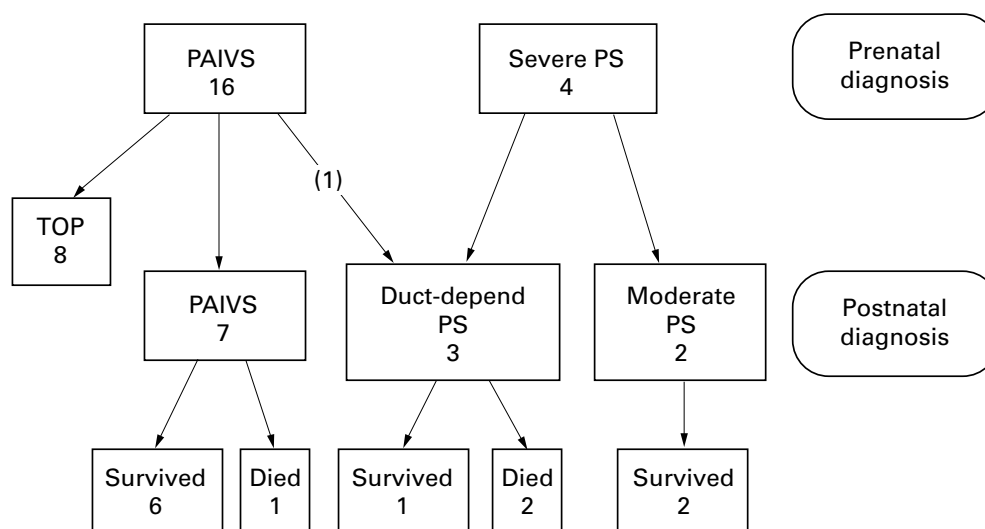
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┌──────────────┐        ┌──────────────┐              ╭──────────────╮
│    PAIVS     │        │  Severe PS   │              │   Prenatal   │
│      16      │        │      4       │              │  diagnosis   │
└──────────────┘        └──────────────┘              ╰──────────────╯
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Figure 1 shows a flow diagram with boxes: PAIVS 16, Severe PS 4, Prenatal diagnosis; TOP 8, PAIVS 7, Duct-depend PS 3, Moderate PS 2, Postnatal diagnosis; (1); Survived 6, Died 1, Survived 1, Died 2, Survived 2.

Figure 1   Prenatal and postnatal diagnosis, and outcome of 20 cases with prenatal diagnosis of right ventricular outflow tract obstructive lesion and intact ventricular septum. PAIVS, pulmonary atresia and intact ventricular septum; PS, pulmonary stenosis; TOP, termination of pregnancy; duct-depend, ductus arteriosus dependent.

straight course before joining at the bifurcation of the pulmonary trunk.

Results
Twenty fetuses were included in the study (table 1, fig 1). Gestational age at presentation was a median of 28.5 weeks, with a range of 17–38 weeks. All 15 fetuses with PAIVS (cases 1–15) and two with pulmonary stenoses (cases 16 and 17) were referred for fetal echocardiography following an abnormal screening ultrasound examination suspicious for fetal cardiac disease. The three other cases of pulmonary stenoses (cases 18–20) were referred for fetal echocardiography as high risk pregnancies. A family history was found in two of the five pulmonary stenoses cases. There were two cases of twin pregnancy (cases 4 and 18). During this study period, while 15 fetuses with PAIVS were detected, 60 other cases with PAIVS were diagnosed postnatally in our institution.

ACCURACY OF THE PRENATAL DIAGNOSIS
Of the 20 cases of right ventricular outflow tract obstruction, 17 had confirmation of their diagnosis (fig 1). Five had confirmation of PAIVS by postmortem examination following termination of pregnancy, and 12 with PAIVS (n = 8) or pulmonary stenosis (n = 4) had postnatal confirmation by echocardiography or angiography. Neither postnatal nor postmortem confirmation was available in three.

At the prenatal examination, 16 cases had no visible flow through the pulmonary valve and had retrograde flow in the ductus arteriosus, and were diagnosed as PAIVS. Of these, 13 had a postnatal investigation, which confirmed the diagnosis in 12, with one other having critical pulmonary stenosis as diagnosed by a right ventriculogram (case 16). Three other cases were terminated and had no confirmation of the diagnosis. Of these 16, three had mild and three moderate tricuspid valve regurgitation. There was no false negative diagnosis in this series, and one false positive in a case who postnatally had critical pulmonary stenosis.

Four cases were diagnosed as severe pulmonary stenosis prenatally with the presence of antegrade turbulent flow through the pulmonary valve, decreased right ventricular function, and antegrade flow in the ductus arteriosus. At birth, two of these four had critical pulmonary stenosis with ductus dependent pulmonary circulation (cases 17 and 18), while the other two had pulmonary stenosis that was
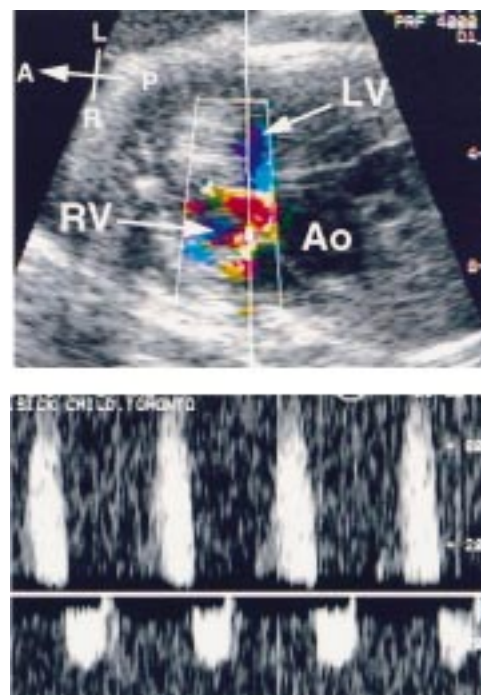

Figure 2   Prenatal echocardiograms in case 3 at 34 weeks' gestation. The upper panel is a cross sectional and colour Doppler image at a transverse section immediately superior to the five chamber view. It demonstrates a turbulent colour signal at the right ventricle (RV) free wall and the ventricular septum during systole, and blow of this turbulent flow into the aorta (AO). In the lower panel, pulsed Doppler interrogation in a right coronary artery (as shown in the upper panel) shows systolic high velocity retrograde flow and diastolic low velocity antegrade flow.
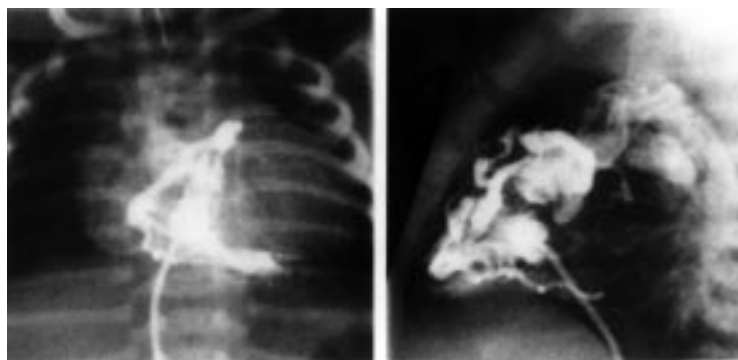
Figure 3 A postnatal right ventriculogram for case 3 reveals an aneurysmal dilated right coronary artery and large sinusoids in the ventricular septum.

not ductus dependent. Of this group one case had mild and one moderate tricuspid regurgitation.

POSTNATAL OUTCOMES

Eight of the 15 patients with PAIVS were diagnosed before the 24th week of gestation; all eight pregnancies were electively terminated (table 1, fig 1). The remaining seven cases of PAIVS and the five with pulmonary stenosis survived to term. In three of these 12 live births (cases 8, 16, and 18) it was decided to undertake no active treatment because of associated non-cardiac lesions, and all three died in the neonatal period.

Of the nine infants who lived, five of the six with PAIVS are on a single ventricle repair track because of severe right ventricular hypoplasia (post-Fontan procedure in one child and post-bidirectional cavopulmonary shunt in four). The sixth PAIVS case awaits biventricular repair (with post-right modified Blalock-Taussig shunt and right ventricular decompres-
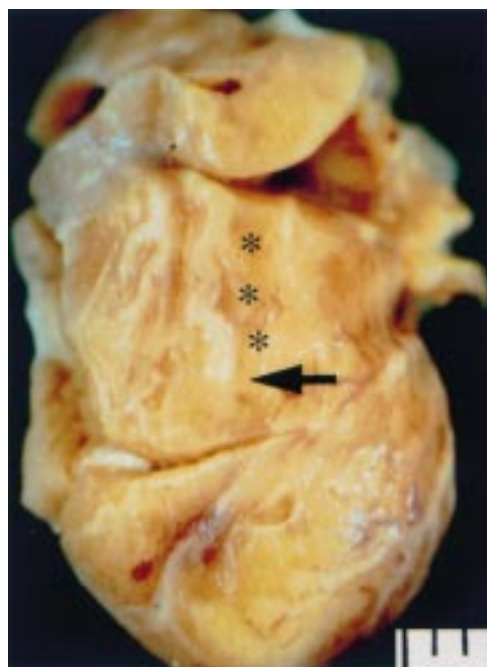

Figure 4 Necropsy of case 5 reveals a mildly dilated tortuous right coronary artery (★), which suddenly disappears from the right ventricular surface and goes into the myocardium (arrow), suggesting the presence of ventriculocoronary connections (scale—mm).

sion). All three surviving cases with pulmonary stenosis have undergone successful valvuloplasty—two balloon and one surgical valvuloplasty. Postnatal follow up in these nine cases had a median length of 2.2 years (range 1.4–7.4 years).

ASSOCIATED ABNORMALITIES

Of the 20 fetuses, three had an associated extracardiac abnormality. Associated Dandy-Walker syndrome was found prenatally in case 8. One aborted fetus (case 13) was found to be dysmorphic at necropsy, which had not been observed by means of prenatal ultrasound. Chromosome 4p–deletion was found in another child (case 16) after birth.

PRENATAL DETECTION OF VENTRICULOCORONARY CONNECTIONS

Of the 20 cases diagnosed prenatally with PAIVS or pulmonary stenosis, 13 (cases 2–7, 9, 10, 14, 15, 17, 18, and 20) were diagnosed after February 1994 and had actively assessed the presence of ventriculocoronary connections (table 1). Before that time, a systematic evaluation of the aortic root and right ventricular myocardium was not performed.

Obvious connections (typical ventriculocoronary connections flow patterns) were observed prenatally by colour and pulsed Doppler in five of 13 cases, and all these five had PAIVS. This typical turbulent flow in the coronary artery was best seen at the transverse section of the fetal heart and immediately superior to the five chamber view (fig 2). In two cases, postnatal right ventriculograms confirmed the presence of large connections (fig 3). The other three cases were pregnancies terminated with no opportunity for ventriculogram, but one necropsy (case 5) revealed a moderately dilated tortuous right coronary artery (fig 4), suggesting the presence of large ventriculocoronary connections. The other cases (cases 2 and 6) had no confirmation.

Suspicious connections were diagnosed prenatally in two cases, both with PAIVS; both underwent a postnatal right ventriculogram, which revealed small ventriculocoronary connections (case 7) and absence of notable ventriculocoronary connections (case 10). One prenatal diagnosis had been falsely positive. Upon retrospective review of the videotape of the fetal echocardiogram in case 10, an artificial turbulent colour signal in the epicardial surface of the myocardium was identified, but no turbulent flow came from the endomyocardium.

Ventriculocoronary connections in six cases (three with PAIVS and three with pulmonary stenosis) were diagnosed as absent (having no notable connections). Three (one PAIVS, two pulmonary stenosis) were confirmed by right ventriculogram; the other three did not have confirmation.

HEART CHAMBER AND VESSEL SIZE

Of the 15 cases of PAIVS, five had both prenatal and postnatal echocardiographic measurements, and the other 10 had only prenatal measurements. All five cases of pulmonary
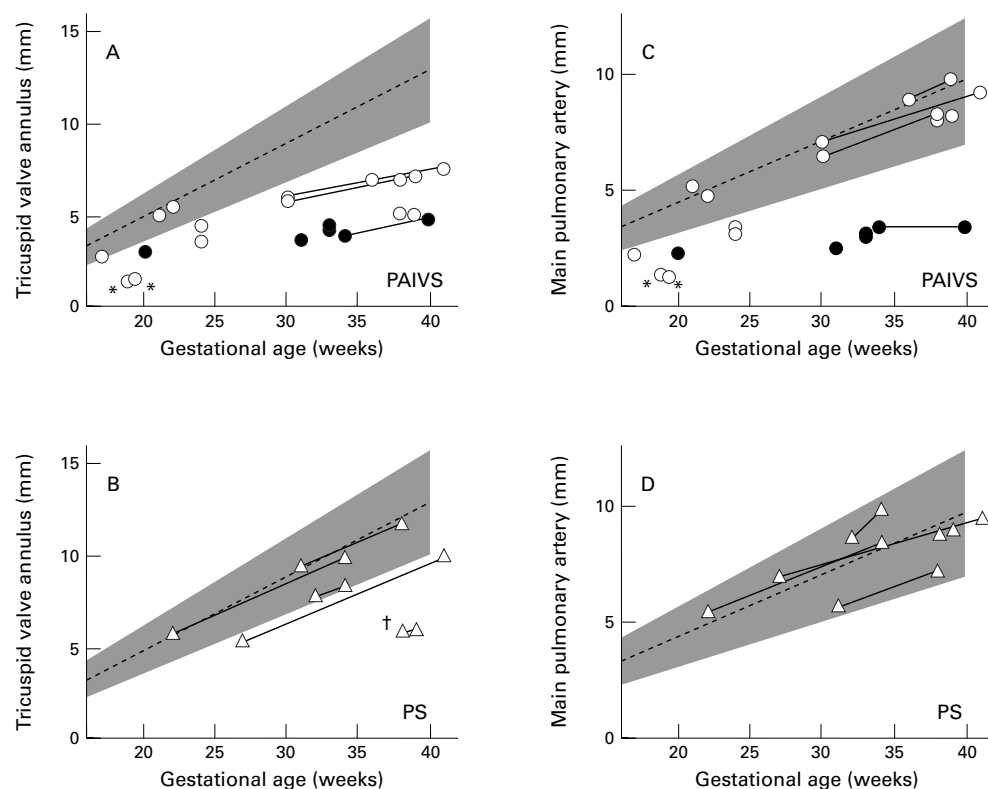
*Figure 5 Diameter of the tricuspid valve annulus and the main pulmonary artery during gestation: diameter of the tricuspid valve annulus in cases with PAIVS (A), and with pulmonary stenosis (B); diameter of the main pulmonary artery in cases with PAIVS (C), and with pulmonary stenosis (D). The 5th and 95th centiles measured in normal fetuses are shown by the grey area. Postnatal measurements at birth are plotted at the gestational age at birth. Points joined together represent prenatal and postnatal measurements obtained from the same case. Black circles represent cases who had confirmation of presence of large ventriculocoronary connections. *Cases with obvious Doppler flow pattern of ventriculocoronary connections detected by prenatal echocardiography but unconfirmed (cases 2 and 6); †case 16 with small ventriculocoronary connections.*

stenosis had both prenatal and postnatal echocardiographic measurements.

Prenatally, the diameters of the tricuspid valve annulus were less than −2 SD in 12 of 15 fetuses with PAIVS and in two of five fetuses with pulmonary stenosis (fig 5A, B), in cases 2 and 6 the measurement being −2 SD at 19 weeks' gestation. Although with our small numbers a significant difference between those with PAIVS and pulmonary stenosis could not be demonstrated, cases with PAIVS seemed to have more severe hypoplasia of the tricuspid valve annulus than those with pulmonary stenosis. Only one fetus with pulmonary stenosis (case 16), who had retrograde flow in the ductus arteriosus and small ventriculocoronary connections, had severe hypoplasia. In all five with PAIVS who had serial measurements, the degree of hypoplasia of the tricuspid annulus assessed by z score became more pronounced at birth.

The diameters of the main pulmonary artery were less than −2 SD in nine of 15 fetuses with PAIVS, especially in all four who had large ventriculocoronary connections (fig 5C). All five fetuses with pulmonary stenosis, however, had the main pulmonary artery within normal range (fig 5D). The postnatal measurement in a case with PAIVS and large ventriculocoronary connections (case 1) demonstrated that its diameter had only minimally increased, diverging from the centiles during gestation, whereas the diameters remained within the normal range in all four cases with PAIVS without

ventriculocoronary connections and all five with pulmonary stenosis.

Measurements of the left heart (mitral valve annulus diameter and ascending aorta diameter) were normal or slightly larger than normal throughout gestation.

The shape of the ductus arteriosus was identified in 18 cases (table 1). A sigmoid shaped ductus arteriosus was found in significantly more cases with PAIVS associated with large ventriculocoronary connections (one of these cases had a prenatal diagnosis of obvious ventriculocoronary connections but lacked postmortem confirmation) than those with PAIVS without large ventriculocoronary connections or those with pulmonary stenosis (p = 0.008) by Fisher's test.

Discussion

Right ventricular outflow tract obstruction (pulmonary atresia or pulmonary stenosis) with intact ventricular septum is a complex disorder. It has a wide anatomical and clinical spectrum involving all components of the right ventricle, with the potential for persistence of primitive ventriculocoronary connections. Providing an accurate description of its morphological variants both prenatal and postnatally is more than just an academic exercise. Several long term follow up studies of patients with PAIVS have clearly demonstrated that management and prognosis are linked to the degree of

right ventricular hypoplasia, as well as the presence or absence of ventriculocoronary connections.[2 15–18]

ACCURACY OF PRENATAL ECHOCARDIOGRAPHY

Our study confirms the accuracy of prenatal diagnosis of pulmonary atresia and severe stenosis with intact ventricular septum. For the prenatal diagnosis of PAIVS, there were no false negatives, and one false positive. Differential diagnosis between pulmonary atresia and critical stenosis is still difficult.[7] Although some cases of critical stenosis can progress to pulmonary atresia in utero,[8] differentiation of these two groups may not affect their prenatal and postnatal management because of its similar strategy. Some stenotic lesions have been reported to progress during the prenatal period.[6 19] In fact, in our study two cases with a postnatal diagnosis of ductal dependent pulmonary stenosis had demonstrable antegrade ductal flow prenatally. Therefore, reassessment in later gestation before a decision is made about early postnatal management is important.

Importantly, this study demonstrates that ventriculocoronary connections can be identified with current ultrasound equipment even as early as 19 weeks' gestation. This lesion was documented at 30 weeks' gestation in one case report.[9] Prenatal detection of other coronary abnormalities has also been reported. Abnormal coronary flow has been demonstrated as early as 24 weeks in a fetus with intrauterine growth retardation.[20] Sharland and colleagues identified an isolated coronary artery fistula at 20 weeks in one fetus.[21] Ventriculocoronary connections can be identified in cases of PAIVS because of their haemodynamic feature. A high pressure head from the right ventricle to the aorta in systole, and from the aorta to the right ventricle in diastole, results in a relatively high velocity to and fro flow in the coronary artery, which can readily be detected by colour flow Doppler. The coronary artery, which is also frequently dilated, may be imaged in some cases with high resolution two dimensional imaging.

PRENATAL RIGHT HEART DIMENSION

At the early stage of fetal development, primitive right ventricular coronary artery connections still exist, which regress in the normal heart which has a patent pulmonary outflow tract. The presence of right heart hypoplasia is believed to be related to abnormal right heart flow; it would then follow that the earlier the insult, the greater the degree of right heart hypoplasia. Similarly, there is a greater likelihood that the primitive ventriculocoronary connections would persist in those with severe right ventricular hypoplasia.

We had excluded those with a large right ventricle and severe tricuspid regurgitation, because prognosis and management during the prenatal period is probably very different.[5 7 8 22 23] Although we cannot exclude some effect of tricuspid regurgitation on right ventricle and tricuspid valve growth in our patients, excluding those with notable tricuspid

valve pathology made this a reasonably homogeneous group.

We demonstrated that in fetuses with right ventricular outflow tract obstruction and intact ventricular septum, the right heart is small in the second and third trimester and that this hypoplasia may progress during the remaining gestation. These observations are similar to results from a study of human fetuses with left ventricular hypoplasia in association with left ventricular outflow obstruction.[14] However, from our study it is difficult to assess precisely the relation between right ventricular growth and either the presence of ventriculocoronary connections or the severity of pulmonary outflow obstruction, because of the small number of subjects in each group.

Despite this, our study supports the theory that a hypoplastic right ventricle and ventriculocoronary connections may be associated with a right ventricular outflow tract obstruction occurring very early in gestation. Most of the fetuses in our study with large ventriculocoronary connections also had a sigmoid ductus arteriosus and a severely hypoplastic right ventricular outflow tract. Sigmoid ductus is thought to signify early onset of right ventricular outflow tract obstruction during heart development, according to findings from postnatal cases of pulmonary atresia with or without a ventricular septal defect.[24 25] In contrast, we observed normal ductus arteriosus morphology in fetuses with PAIVS and pulmonary stenosis without notable ventriculocoronary connections in which the right ventricular outflow tract was nearer to normal.

Main pulmonary artery size seemed to be within normal limits throughout gestation in the group having PAIVS without ventriculocoronary connections, even though the pulmonary valve was already closed, with no antegrade flow. This finding cannot be explained by Kutsche and Van Mierop's theory that normal antegrade blood flow during much or most of the prenatal period, in combination with poststenotic dilation, result in a main pulmonary artery of normal or even larger size.[25] Our findings also differ from those observed in left heart outflow obstruction,[14] in which all fetuses with aortic valve atresia or aortic stenosis had significantly reduced aortic growth. We hypothesise that the shape of the ductus arteriosus has an important effect on the growth of the main pulmonary artery. We found all fetuses with a sigmoid ductus arteriosus to have a smaller main pulmonary artery, whereas all those with a normally shaped, straight ductus arteriosus had a normal main pulmonary artery. One could speculate that less retrograde ductal flow in those with a sigmoid duct, in conjunction with an earlier developmental lesion, might explain the difference in pulmonary artery size between cases with and without ventriculocoronary connections.

PRENATAL AND POSTNATAL MANAGEMENT

Although no intrauterine death occurred in our series, our population had an obvious bias. First, we excluded fetuses with tricuspid valve dysplasia and Ebstein's malformation, who

may have associated anatomical pulmonary atresia or pulmonary stenosis and who have a poor prognosis because of associated tricuspid regurgitation.[22] [23] Second, we cannot know the intrauterine course of the seven terminated cases diagnosed before the 24th week of gestation, all of whom had serious right ventricular hypoplasia. Despite this, we can gain some insight into in utero survival from published data. Allan and associates reported that 9% of fetuses with PAIVS or severe pulmonary stenosis in the absence of notable tricuspid valve regurgitation died in utero.[8]

Notably, 15% of our 20 fetuses had either dysmorphic features or a chromosomal abnormality. Allan and associates also reported that 20% of fetuses with PAIVS or pulmonary stenosis were found to have chromosomal abnormalities.[8] Like other forms of congenital heart disease, chromosomal analysis is a very important component of family counselling. In the antenatal diagnosis of right ventricular outflow tract obstruction, a systemic obstetrical ultrasound examination and chromosome analysis should be considered.

Although our study showed no postnatal death in the patients with active treatment regardless of presence or absence of ventriculocoronary connections, this result cannot be interpreted as representing an improved outcome in the patients with prenatal diagnosis of PAIVS. Our patient group is biased because of the number of terminations, a short term follow up, and only a few postnatal cases with ventriculocoronary connections. Hence, better postnatal outcome in our study cannot be compared to several previous large series.[2–4] Further studies with large numbers of cases without the influence of termination would be necessary to investigate the advantage of prenatal detection of PAIVS and ventriculocoronary connections on postnatal outcome.

What, therefore, are the implications of this study? We have shown that an accurate delineation of the spectrum of PAIVS is possible using fetal echocardiography, including the detection of large ventriculocoronary connections. This latter information is vital, as several studies demonstrated that the postnatal prognosis of this lesion is directly linked to the presence or absence of ventriculocoronary connections of note. Interruption or stenosis of the coronary artery, which has a profound effect on postnatal management,[2] [3] [17] cannot be determined by prenatal cardiac ultrasound. Nevertheless, the presence of ventriculocoronary connections with or without right ventricular dependent coronary circulation is a significant risk factor for poor prognosis.[2] [18] [26] Hence, diagnosing ventriculocoronary connections in utero is important to family counselling. We found that fetuses with a right ventricular outflow tract obstructive lesion with ventriculocoronary connections have a greater degree of right ventricle and main pulmonary artery hypoplasia, both of which affect outcome. We also suggested that worse growth of the right ventricle and main pulmonary artery in utero might be associated with early onset of right ventricular outflow tract obstruction, suggested by the presence of ventriculocoronary connections and a sigmoid shaped ductus arteriosus. These data can therefore be used in the counselling process for families identified as having a fetus with this anomaly, recognising that data regarding improved outcome are not available at present with a prenatal diagnosis. Prospective serial study of affected fetuses will provide further information on the natural history after birth.

In general, only cases with outflow tract obstruction of consequence can be diagnosed prenatally, and we recognise that these observations most likely do not apply to milder forms of obstruction.

CONCLUSIONS

Current high resolution ultrasound provides accurate information for prenatal diagnosis of right ventricular outflow obstruction and ventriculocoronary connections. In this study, the size of the main pulmonary artery and the tricuspid valve annulus, and the shape of the ductus arteriosus, seemed to be associated with the presence of ventriculocoronary connections. Prenatal detection of this constellation of abnormalities may aid in family counselling and decisions on postnatal management.

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